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When a lymphatic malformation determines a bowel volvulus: Are clinical status and images always reliable?

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ABSTRACT

INTRODUCTION: An acute abdomen in the form of small-bowel volvulus could be a presentation of a lymphatic malformation in childhood.
CASE PRESENTATION: A 5 year old male was admitted to our Institute for an acute abdomen. Clinical aspects and radiological images were not specific for a certain diagnosis. Laparotomy revealed a big soft mass, with a milky content, completely involving about 50 cm of ileus with a partial volvulus of the intestinal loop. A complete mass excision and also a bowel involved resection were performed.
After a histological examination, a lymphatic malformation was diagnosed.
DISCUSSION: The diagnosis of a mesenteric lymphatic malformation could be intraoperative and a complete resection should be the treatment of choice. Sometimes it could be necessary to perform an involved bowel tract resection in the case of volvolus with ischemia.
CONCLUSIONS: Paediatricians and surgeons should bare in mind that an intrabdominal lymphatic malformation may present as a nonspecific an acute abdomen caused by a bowel volvolus and diagnosis may not be so simple preoperatively.

1. Introduction

Lymphatic malformations (LM) can be encountered in the viscera in about 10% of all cases and are incline to be lesions in childhood and infancy because they are probably due to a congenital malformation of the lymphatic vessels. The most frequent intrabdominal site is the small intestine mesentery (70.5%) [1,2].

Clinical signs and symptoms are not always specific, and despite using US, MRI, and TC diagnosis could be intraoperative [2,3].

We propose a case of an intraoperative diagnosis of a mesenteric macrocystic LM presenting in the form of a small-bowel volvulus with an acute abdomen.

2. Case report

A 5 year old male was admitted to our Institute for an acute abdomen, with an abrupt abdominal pain.

The pain had started about 8 h earlier in the morning and symptoms had worsened in the last four hours.

The pain was unresponsive to oral analgesic therapy.

Some vomiting was reported in the previous hours.

No diarrhoea or fever was associated.

No previous episodes of abdominal pain were reported.

The patient had not lost or gained weight in the last month.

The abdomen was very distended, diffusely very painful and peristalsis was poor.

Parents had reported a distended painless abdomen appeared for several weeks.

Vital signs were stable.

A normal white blood cells count and a normal C-reactive protein level were documented.

Ultrasonography (US) showed a widespread dilated fluid-filled bowel and a mesogastric image of multiple small anechoic areolas of around one centimeter placed in a thickening and hypervascularised mesenteric adipose tissue of uncertain significance (Fig. 1).

Abbreviations: CT, computed tomography; US, ultrasonography; MRI, magnetic resonance; LM, lymphatic malformation.

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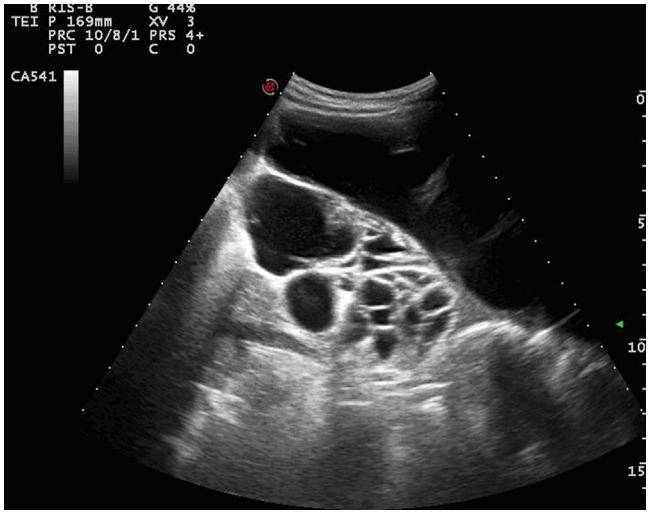

Fig. 1. US showed a widespread dilated fluid-filled bowel and a mesogastric image of multiple anechoic areolas.

Fig. 2. CT highlighted the important distension of a central abdominal intestinal loop with air-fluid levels and a convolute aspect, suspect for intestinal volvolus.

Abdominal radiography and CT highlighted the important distension of a central abdominal intestinal loop with air-fluid levels, with a convolute aspect, suspect for intestinal volvulus originating from the small bowel (Fig. 2).

Suspecting a bowel volvolus, we decided to perform a laparotomy revealing, after the opening of peritoneum, a very big soft mass, with a milky content, completely involving about 50 centimetres of the ileus (130 cm from the Treitz ligament) and originating from mesenteric adipose tissue. The mass determines a partial volvulus of the intestinal loop, without significant ischemia. Several voluminous lymph nodes were attached to the mass (Fig. 3). After solving the volvulus, a completed abdominal mass excision was conducted performing a 50 cm bowel involved resection as well (Fig. 4). An end to end anastomosis was performed.

Triple antibiothic therapy with ampicillin/sulbactam (50 mg/kg 3 times a day), metronidazole (7,5 mg/kg 3 times a day) and tobramycin (5 mg/kg once a day) were started intraoperatively.

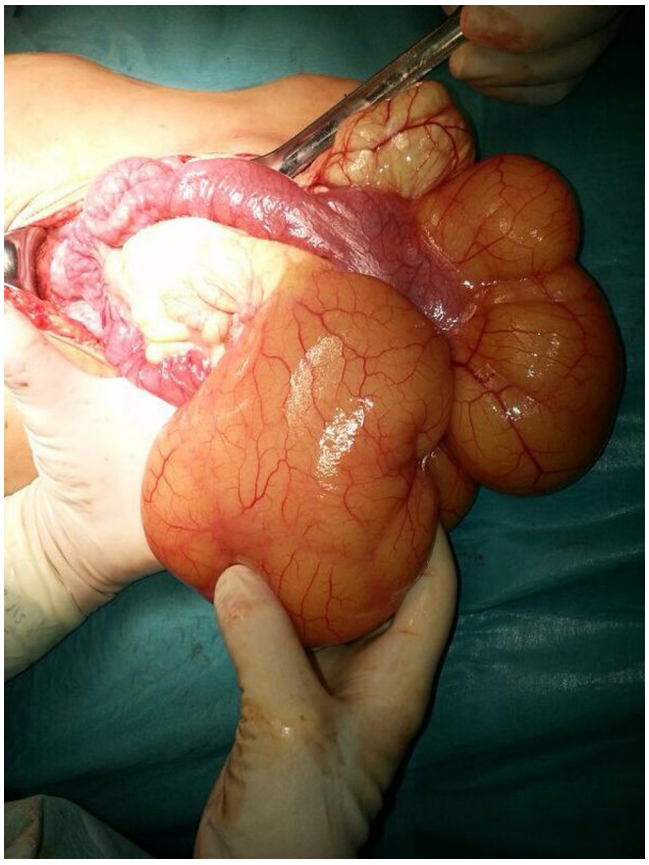

Fig. 3. A big soft mass, with a milky content, completely involving about 50 centimetres of ileus was revealed after a laparotomy.

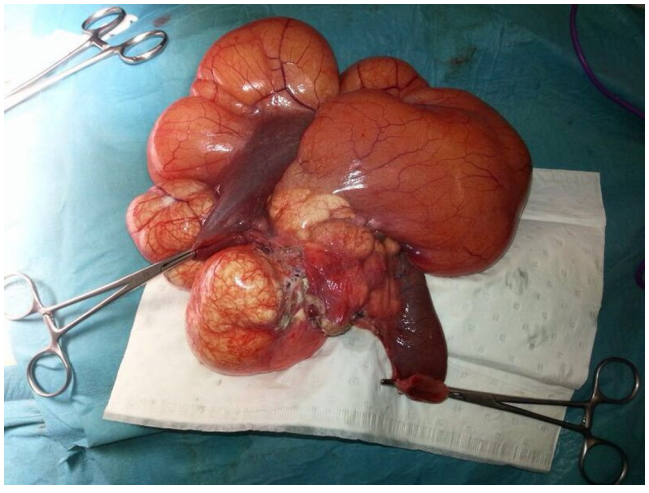

Fig. 4. A completed abdominal mass excision was done including a 50 centimetres bowel resection.

Postoperative analgesia was administered via an elastomeric pump to provide adequate pain control.

Nasogastric tube and vesical catheter were removed on 2nd postoperative day.

Solid feeding was started on 3rd postoperative day after bowel movements restartied.

No post operative complications were observed and the child was discharge at day 6.

The histological examination showed the lesion was composed by numerous dilated, irregularly shaped lymphatic channels and a

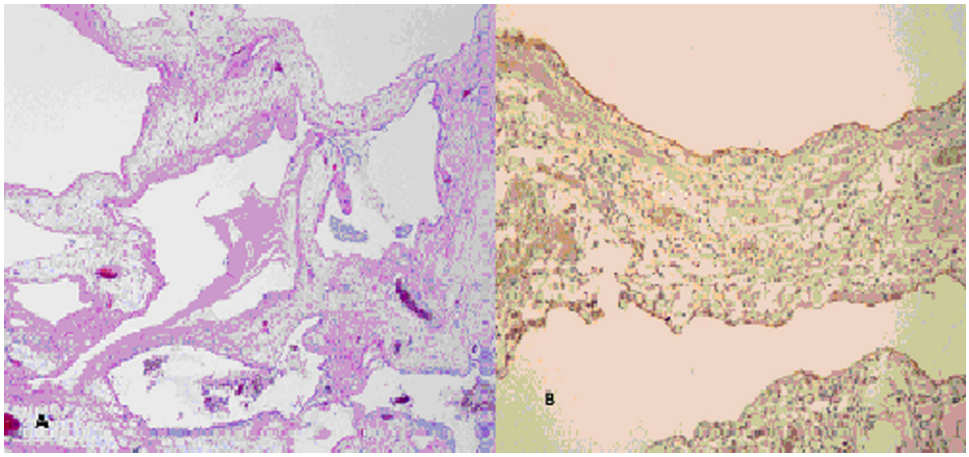

Fig. 5. A – Hematoxilin and Eosin, x2.5 The lesion is composed by numerous dilated, irregularly shaped lymphatic channels. B – The vascular channels highlighted with D2-40 immunostaining.

macrocystic lymphatic malformation was diagnosed. The channels were highlighted with D2-40 immunostaining (Fig. 5).

No residual disease was detected at MRI performed 4 months after surgery.

3. Discussion

Small-bowel volvulus with an acute abdomen could be the presentation of a mesenteric LM that typically presents as a chronic abdominal distension with palpable abdominal mass and, in some cases, a progressive complete or partial bowel occlusion [4,5]. US and MRI are considered the most appropriate radiodiagnostic instruments to evaluate LMs [6], and T2 weighted MRI is the gold standard diagnosis in elective treatment. We used CT in emergency suspecting a bowel volvolus. However, this imaging modality may not have a diagnostic specificity, and so diagnosis of a mesentheric LM could be intraoperative only. At our Institute, we have also recently introduced MRI as diagnostic imaging modality in selected cases of abdominal emergency to improve diagnostic accuracy.

In literature, LMs are generically distinguished as macrocystic (>2 cm) and microcystic (<2 cm). In elective, surgery it is generally not recommended, as the first therapy, for intraperitoneal macrocystic lesions, and drainage of the macrocyst cavity under US guidance puncture with a following sclerosant (such as Docycycline) injection are the gold standard procedures and multiple treatment sessions may be necessary [1].

However, surgery could be required in abdominal emergency suspecting a bowel volvolus or in case of macrocyst LM puncture complications such as cystic wall rupture, leakage, infection or adjacent organ injury [1]. Resection of a mesenteric LM may sometimes require resecting an involved bowel tract because it is often strongly attached to the bowel vascularization and, in case of volvulus, a several bowel ischemia may occur [7].

If surgery is the choice, a complete resection of the cyst should be the treatment of choice, while a simple aspiration or an incomplete resection are associated with a high recurrence rate [4,5]. Moreover, some studies have reported a malignant change, so a complete resection should be performed in all cases [4,5].

In conclusion, the paediatricians and surgeons should bear in mind that macrocystic intraperitoneal LM, in some cases, may present as an acute abdomen caused by a bowel volvolus and diagnosis may not be so simple preoperatively.

Conflicts of interest

There is no statement of any potential conflict of interest, real or perceived.

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Ethical approval

This study did not need of any Ethical Approval, not being a research study.

Consent

"Written informed consent was obtained from the patient".

Author contribution

Edoardo Guida and Maria Antonietta Lembo wrote the first draft of the manuscript. Massimo Di Grazia contributed to data collection. Elisabetta Cattaruzzi contributed to radiological data collection Rossana Bussani contributed to immunohistological data collection. Waifro Rigamonti supervised this study.

Guarantor

Edoardo Guida, MD.

References

[1] N. Nassiri, J. Thomas, N.C. Cirillo-Penn, Evaluation and management of peripheral venous and lymphatic malformations, J. Vasc. Surg. Venous Lymphat. Disord. 4 (April (6)) (2016) 257–265.
[2] V.B. Weeda, K.A. Booij, D.C. Aronson, Mesenteric cystic lymphangioma: a congenital and an acquired anomaly? Two cases and a review of the literature, J. Pediatr. Surg. 43 (June (6)) (2008) 1206–1208.
[3] H. Tsukada, K. Takaori, S. Ishiguro, et al., Giant cystic lymphangioma of the small bowel mesentery: report of a case, Surg. Today 32 (8) (2002) 734–737.

[4] S. Talukdar, S. Alagaratnam, A. Sinha, et al., Giant cystic lymphangioma in childhood: a rare differential for the acute abdomen, BMJ Case Rep. (2011), July 7.
[5] W. Suthiwartnarueput, S. Kiatipunsodsai, A. Kwankua, et al., Lymphangioma of the small bowel mesentery: a case report and review of the literature, World J. Gastroenterol. 18 (43) (2012) 6328–6332, November 21.
[6] O. Konen, V. Rathaus, E. Dlugy, et al., Childhood abdominal cystic lymphangioma, Pediatr. Radiol. 32 (February (2)) (2002) 88–94.
[7] V.B. Weeda, K.A. Booij, D.C. Aronson, Mesenteric cystic lymphangioma: a congenital and an acquired anomaly? Two cases and a review of the literature, J. Pediatr. Surg. 43 (June (6)) (2008) 1206–1208.